



SEQUENCE LISTING

<110> Biofrontera Pharmaceuticals AG
Lubbert, Hermann

<120> TRANSGENIC ANIMAL MODEL FOR
NEURODEGENERATIVE DISEASES

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<140> 09/830,703
<141> 2001-04-26

<150> EP 99116766.9
<151> 1999-08-30

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Pro Val Asp Ser Val Gly Leu Ala Val Ile Leu Asp Thr Asp Ser Lys
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35 40 45
Glu Leu Pro Asn His Leu Thr Val Gln Leu Asn Pro Pro Thr Thr Ala
50 55 60
Phe Ser Ser Thr Ala Lys Ala Pro Ala Thr Arg Ser Ser Leu Glu Ser
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50 55 60
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Ser Asp Lys Asp Thr Ser Val Ala Leu Asn Leu Ile Thr Ser Asn Arg

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 Trp Glu Ser Arg Ser Leu Thr Arg Val Asp Leu Ser Ser His Thr Leu
 100 105 110
 Pro Val Asp Ser Val Gly Leu Ala Val Ile Leu Asp Thr Asp Ser Lys
 115 120 125
 Arg Asp Ser Glu Ala Ala Arg Gly Pro Gly Pro Ile Leu Leu Gly Arg
 130 135 140
 Cys Leu Asn Ser Lys Pro Asp Glu Trp
 145 150

<210> 25
 <211> 194
 <212> PRT
 <213> Mus musculus

<400> 25
 Met Ile Val Phe Val Arg Phe Asn Ser Ser Tyr Gly Phe Pro Val Glu
 1 5 10 15
 Val Asp Ser Asp Thr Ser Ile Leu Gln Leu Lys Glu Val Val Ala Lys
 20 25 30
 Arg Gln Gly Val Pro Ala Asp Gln Leu Arg Val Ile Phe Ala Gly Lys
 35 40 45
 Glu Leu Pro Asn His Leu Thr Val Gln Asn Cys Asp Leu Glu Gln Gln
 50 55 60
 Ser Ile Val His Ile Val Gln Arg Pro Arg Arg Arg Ser His Glu Thr
 65 70 75 80
 Asn Ala Ser Gly Gly Asp Glu Pro Gln Ser Thr Ser Glu Gly Ser Ile
 85 90 95
 Trp Glu Ser Arg Ser Leu Thr Arg Val Asp Leu Ser Ser His Thr Leu
 100 105 110
 Pro Val Asp Ser Val Gly Leu Ala Val Ile Leu Asp Thr Asp Ser Lys
 115 120 125
 Arg Asp Ser Glu Ala Ala Arg Gly Pro Val Lys Pro Thr Tyr Asn Ser
 130 135 140
 Phe Phe Ile Tyr Cys Lys Gly Pro Cys His Lys Val Gln Pro Gly Lys
 145 150 155 160
 Leu Arg Val Gln Cys Gly Thr Cys Lys Gln Ala Thr Leu Thr Leu Ala
 165 170 175
 Gln Asn Phe Ser Leu Asn Val Glu His Thr Gln Pro Gln Thr Arg Thr
 180 185 190
 Arg Arg

<210> 26
<211> 183
<212> PRT
<213> Mus musculus

<400> 26

Met Ile Val Phe Val Arg Phe Asn Ser Ser Tyr Gly Phe Pro Val Glu
1 5 10 15
Val Asp Ser Asp Thr Ser Ile Leu Gln Leu Lys Glu Val Val Ala Lys
20 25 30
Arg Gln Gly Val Pro Ala Asp Gln Leu Arg Val Ile Phe Ala Gly Lys
35 40 45
Glu Leu Pro Asn His Leu Thr Val Gln Asn Cys Asp Leu Glu Gln Gln
50 55 60
Ser Ile Val His Ile Val Gln Arg Pro Arg Arg Arg Ser His Glu Thr
65 70 75 80
Asn Ala Ser Gly Gly Asp Glu Pro Gln Ser Thr Ser Glu Gly Ser Ile
85 90 95
Trp Glu Ser Arg Ser Leu Thr Arg Val Asp Leu Ser Ser His Thr Leu
100 105 110
Pro Val Asp Ser Val Gly Leu Ala Val Ile Leu Asp Thr Asp Ser Lys
115 120 125
Arg Asp Ser Glu Ala Ala Arg Gly Pro Val Lys Pro Thr Tyr Asn Ser
130 135 140
Phe Phe Ile Tyr Cys Lys Gly Pro Cys His Lys Val Gln Pro Gly Lys
145 150 155 160
Leu Arg Val Gln Cys Gly Thr Cys Lys Gln Ala Thr Leu Thr Leu Ala
165 170 175
Gln Leu Ala Val Pro Thr Pro
180

<210> 27
<211> 296
<212> PRT
<213> Mus musculus

<400> 27

Met Ile Val Phe Val Arg Phe Asn Ser Ser Tyr Gly Phe Pro Val Glu
1 5 10 15
Val Asp Ser Asp Thr Ser Ile Leu Gln Leu Lys Glu Val Val Ala Lys
20 25 30
Arg Gln Gly Val Pro Ala Asp Gln Leu Arg Val Ile Phe Ala Gly Lys
35 40 45
Glu Leu Pro Asn His Leu Thr Val Gln Asn Cys Asp Leu Glu Gln Gln
50 55 60
Ser Ile Val His Ile Val Gln Arg Pro Arg Arg Arg Ser His Glu Thr
65 70 75 80
Asn Ala Ser Gly Gly Asp Glu Pro Gln Ser Thr Ser Glu Gly Ser Ile
85 90 95
Trp Glu Ser Arg Ser Leu Thr Arg Val Asp Leu Ser Ser His Thr Leu
100 105 110
Pro Val Asp Ser Val Gly Leu Ala Val Ile Leu Asp Thr Asp Ser Lys
115 120 125

Arg Asp Ser Glu Ala Ala Arg Gly Pro Val Lys Pro Thr Tyr Asn Ser
130 135 140
Phe Phe Ile Tyr Cys Lys Gly Pro Cys His Lys Val Gln Pro Gly Lys
145 150 155 160
Leu Arg Val Gln Cys Gly Thr Cys Lys Gln Ala Thr Leu Thr Leu Ala
165 170 175
Gln Gly Pro Ser Cys Trp Asp Asp Val Leu Ile Pro Asn Arg Met Ser
180 185 190
Gly Glu Cys Gln Ser Pro Asp Cys Pro Gly Thr Arg Ala Glu Phe Phe
195 200 205
Phe Lys Cys Gly Ala His Pro Thr Ser Asp Lys Asp Thr Ser Val Ala
210 215 220
Leu Asn Leu Ile Thr Ser Asn Arg Arg Ser Ile Pro Cys Ile Ala Cys
225 230 235 240
Thr Asp Val Arg Ser Pro Val Leu Val Phe Gln Cys Asn His Arg His
245 250 255
Val Ile Cys Leu Asp Cys Phe His Leu Tyr Cys Val Thr Arg Leu Asn
260 265 270
Asp Arg Gln Phe Val His Asp Ala Gln Leu Gly Tyr Ser Leu Pro Cys
275 280 285
Val Val Cys Phe Leu Pro Gly Leu
290 295

<210> 28
<211> 37
<212> PRT
<213> Mus musculus

<400> 28
Met Ile Val Phe Val Arg Phe Asn Ser Ser Tyr Gly Phe Pro Val Glu
1 5 10 15
Val Asp Ser Asp Thr Ser Ile Leu Gln Leu Lys Glu Val Val Ala Lys
20 25 30
Arg Arg Gly Ser Ser
35

<210> 29
<211> 53
<212> PRT
<213> Mus musculus

<400> 29
Met Ile Val Phe Val Arg Phe Asn Ser Ser Tyr Gly Phe Pro Val Glu
1 5 10 15
Val Asp Ser Asp Thr Ser Ile Leu Gln Leu Lys Glu Val Val Ala Lys
20 25 30
Arg Gln Gly Val Pro Ala Asp Gln Leu Arg Val Ile Phe Ala Gly Lys
35 40 45
Glu Leu Pro Ile Thr
50

<210> 30

<211> 77
<212> PRT
<213> Mus musculus

<400> 30
Met Ile Val Phe Val Arg Phe Asn Ser Ser Tyr Gly Phe Pro Val Glu
1 5 10 15
Val Asp Ser Asp Thr Ser Ile Leu Gln Leu Lys Glu Val Val Ala Lys
20 25 30
Arg Gln Gly Val Pro Ala Asp Gln Leu Arg Val Ile Phe Ala Gly Lys
35 40 45
Glu Leu Pro Asn His Leu Thr Val Gln Asn Cys Asp Leu Glu Gln Gln
50 55 60
Ser Ile Val His Ile Val Gln Arg Pro Arg Glu Lys Ser
65 70 75

<210> 31
<211> 14
<212> PRT
<213> Mus musculus

<400> 31
Met Ile Val Thr Val Thr Trp Asn Asn Arg Val Leu Tyr Thr
1 5 10

<210> 32
<211> 464
<212> PRT
<213> Mus musculus

<400> 32
Met Ile Val Phe Val Arg Phe Asn Ser Ser Tyr Gly Phe Pro Val Glu
1 5 10 15
Val Asp Ser Asp Thr Ser Ile Leu Gln Leu Lys Glu Val Val Ala Lys
20 25 30
Arg Gln Gly Val Pro Ala Asp Gln Leu Arg Val Ile Phe Ala Gly Lys
35 40 45
Glu Leu Pro Asn His Leu Thr Val Gln Asn Cys Asp Leu Glu Gln Gln
50 55 60
Ser Ile Val His Ile Val Gln Arg Pro Arg Arg Ser His Glu Thr
65 70 75 80
Asn Ala Ser Gly Gly Asp Glu Pro Gln Ser Thr Ser Glu Gly Ser Ile
85 90 95
Trp Glu Ser Arg Ser Leu Thr Arg Val Asp Leu Ser Ser His Thr Leu
100 105 110
Pro Val Asp Ser Val Gly Leu Ala Val Ile Leu Asp Thr Asp Ser Lys
115 120 125
Arg Asp Ser Glu Ala Ala Arg Gly Pro Val Lys Pro Thr Tyr Asn Ser
130 135 140
Phe Phe Ile Tyr Cys Lys Gly Pro Cys His Lys Val Gln Pro Gly Asn
145 150 155 160
Leu Arg Val Gln Cys Gly Thr Cys Lys Gln Ala Thr Leu Thr Leu Ala
165 170 175

Gln Gly Pro Ser Cys Trp Asp Asp Val Leu Ile Pro Asn Arg Met Ser
 180 185 190
 Gly Glu Cys Gln Ser Pro Asp Cys Pro Gly Thr Arg Ala Glu Phe Phe
 195 200 205
 Phe Lys Cys Gly Ala His Pro Thr Ser Asp Lys Asp Thr Ser Val Ala
 210 215 220
 Leu Asn Leu Ile Thr Ser Asn Arg Arg Ser Ile Pro Cys Ile Ala Cys
 225 230 235 240
 Thr Asp Val Arg Ser Pro Val Leu Val Phe Gln Cys Asn His Arg His
 245 250 255
 Val Ile Cys Leu Asp Cys Phe His Leu Tyr Cys Val Thr Arg Leu Asn
 260 265 270
 Asp Arg Gln Phe Val His Asp Ala Gln Leu Gly Tyr Ser Leu Pro Cys
 275 280 285
 Val Ala Gly Cys Pro Asn Ser Leu Ile Lys Glu Leu His His Phe Arg
 290 295 300
 Ile Leu Gly Glu Glu Gln Tyr Thr Arg Tyr Gln Gln Tyr Gly Ala Glu
 305 310 315 320
 Glu Cys Val Leu Gln Met Gly Gly Val Leu Cys Pro Arg Pro Gly Cys
 325 330 335
 Gly Ala Gly Leu Leu Pro Glu Gln Gly Gln Arg Lys Val Thr Cys Glu
 340 345 350
 Gly Gly Asn Gly Leu Gly Cys Gly Phe Val Phe Cys Arg Asp Cys Lys
 355 360 365
 Glu Ala Tyr His Glu Gly Asp Cys Asp Ser Leu Leu Glu Pro Ser Gly
 370 375 380
 Ala Thr Ser Gln Ala Tyr Arg Val Asp Lys Arg Ala Ala Glu Gln Ala
 385 390 395 400
 Arg Trp Glu Glu Ala Ser Lys Glu Thr Ile Lys Lys Thr Thr Lys Pro
 405 410 415
 Cys Pro Arg Cys Asn Val Pro Ile Glu Lys Asn Gly Gly Cys Met His
 420 425 430
 Met Lys Cys Pro Gln Pro Gln Cys Lys Leu Glu Trp Cys Trp Asn Cys
 435 440 445
 Gly Cys Glu Trp Asn Arg Ala Cys Met Gly Asp His Trp Phe Asp Val
 450 455 460

<210> 33
 <211> 464
 <212> PRT
 <213> Mus musculus

<400> 33
 Met Ile Val Phe Val Arg Phe Asn Ser Ser Tyr Gly Phe Pro Val Glu
 1 5 10 15
 Val Asp Ser Asp Thr Ser Ile Leu Gln Leu Lys Glu Val Val Ala Lys
 20 25 30
 Arg Gln Gly Val Pro Ala Asp Gln Leu Arg Val Ile Phe Ala Gly Lys
 35 40 45
 Glu Leu Pro Asn His Leu Thr Val Gln Asn Cys Asp Leu Glu Gln Gln
 50 55 60
 Ser Ile Val His Ile Val Gln Arg Pro Arg Arg Arg Ser His Glu Thr
 65 70 75 80
 Asn Ala Ser Gly Gly Asp Glu Pro Gln Ser Thr Ser Glu Gly Ser Ile

85	90	95
Trp Glu Ser Arg Ser Leu Thr Arg Val Asp	Leu Ser Ser His	Thr Leu
100	105	110
Pro Val Asp Ser Val Gly Leu Ala Val Ile	Leu Asp Thr Asp Ser Lys	
115	120	125
Arg Asp Ser Glu Ala Ala Arg Gly Pro Val Lys	Pro Thr Tyr Asn Ser	
130	135	140
Phe Phe Ile Tyr Cys Lys Gly Pro Cys His Lys	Val Gln Pro Gly Lys	
145	150	155
Leu Arg Val Gln Cys Gly Thr Cys Lys Gln Ala	Thr Leu Thr Leu Ala	
165	170	175
Gln Gly Pro Ser Cys Trp Asp Asp Val Leu Ile	Pro Asn Arg Met Ser	
180	185	190
Gly Glu Cys Gln Ser Pro Asp Cys Pro Gly Thr Arg	Ala Glu Phe Phe	
195	200	205
Phe Lys Cys Gly Ala His Pro Thr Ser Asp Lys Asp	Thr Ser Val Ala	
210	215	220
Leu Asn Leu Ile Thr Ser Asn Arg Arg Ser Ile	Pro Cys Ile Ala Cys	
225	230	235
Thr Asp Val Arg Ser Pro Val Leu Val Phe	Gln Cys Asn His Arg His	
245	250	255
Val Ile Cys Leu Asp Cys Phe His Leu Tyr Cys Val	Thr Arg Leu Asn	
260	265	270
Asp Arg Gln Phe Val His Asp Ala Gln Leu Gly Tyr	Ser Leu Pro Cys	
275	280	285
Val Ala Gly Cys Pro Asn Ser Leu Ile Lys Glu	Leu His His Phe Arg	
290	295	300
Ile Leu Gly Glu Glu Gln Tyr Thr Arg Tyr Gln	Gln Tyr Gly Ala Glu	
305	310	315
Glu Cys Val Leu Gln Met Gly Gly Val Leu Cys Pro	Arg Pro Gly Cys	
325	330	335
Gly Ala Gly Leu Leu Pro Glu Gln Gly Gln Arg	Lys Val Thr Cys Glu	
340	345	350
Gly Gly Asn Gly Leu Gly Cys Gly Phe Val Phe	Cys Arg Asp Cys Lys	
355	360	365
Glu Ala Tyr His Glu Gly Asp Cys Asp Ser Leu	Leu Glu Pro Ser Gly	
370	375	380
Ala Thr Ser Gln Ala Tyr Arg Val Asp Lys Arg	Ala Ala Glu Gln Ala	
385	390	395
Arg Trp Glu Glu Ala Ser Lys Glu Thr Ile	Lys Lys Thr Asn Lys Pro	
405	410	415
Cys Pro Arg Cys Asn Val Pro Ile Glu Lys Asn	Gly Gly Cys Met His	
420	425	430
Met Lys Cys Pro Gln Pro Gln Cys Lys Leu Glu	Trp Cys Trp Asn Cys	
435	440	445
Gly Cys Glu Trp Asn Arg Ala Cys Met Gly Asp	His Trp Phe Asp Val	
450	455	460

<210> 34
<211> 451
<212> PRT
<213> Mus musculus

<400> 34

Met Ile Val Phe Val Arg Phe Asn Ser Ser Tyr Gly Phe Pro Val Glu
1 5 10 15
Val Asp Ser Asp Thr Ser Ile Leu Gln Leu Lys Glu Val Val Ala Lys
20 25 30
Arg Gln Gly Val Pro Ala Asp Gln Leu Arg Val Ile Phe Ala Gly Lys
35 40 45
Glu Leu Pro Asn His Leu Thr Val Gln Asn Cys Asp Leu Glu Gln Gln
50 55 60
Ser Ile Val His Ile Val Gln Arg Pro Arg Arg Arg Ser His Glu Thr
65 70 75 80
Asn Ala Ser Gly Gly Asp Glu Pro Gln Ser Thr Ser Glu Gly Ser Ile
85 90 95
Trp Glu Ser Arg Ser Leu Thr Arg Val Asp Leu Ser Ser His Thr Leu
100 105 110
Pro Val Asp Ser Val Gly Leu Ala Val Ile Leu Asp Thr Asp Ser Lys
115 120 125
Arg Asp Ser Glu Ala Ala Arg Gly Pro Val Lys Pro Thr Tyr Asn Ser
130 135 140
Phe Phe Ile Tyr Cys Lys Gly Pro Cys His Lys Val Gln Pro Gly Lys
145 150 155 160
Leu Arg Val Gln Cys Gly Thr Cys Lys Gln Ala Thr Leu Thr Leu Ala
165 170 175
Gln Gly Pro Ser Cys Trp Asp Asp Val Leu Ile Pro Asn Arg Met Ser
180 185 190
Gly Glu Cys Gln Ser Pro Asp Cys Pro Gly Thr Arg Ala Glu Phe Phe
195 200 205
Phe Lys Cys Gly Ala His Pro Thr Ser Asp Lys Asp Thr Ser Val Ala
210 215 220
Leu Asn Leu Ile Thr Ser Asn Arg Arg Ser Ile Pro Cys Ile Ala Cys
225 230 235 240
Thr Asp Val Arg Ser Pro Val Leu Val Phe Gln Cys Asn His Arg His
245 250 255
Val Ile Cys Leu Asp Cys Phe His Leu Tyr Cys Val Thr Arg Leu Asn
260 265 270
Asp Arg Gln Phe Val His Asp Ala Gln Leu Gly Tyr Ser Leu Pro Cys
275 280 285
Val Ala Gly Cys Pro Asn Ser Leu Ile Lys Glu Leu His His Phe Arg
290 295 300
Ile Leu Gly Glu Glu Gln Tyr Thr Arg Tyr Gln Gln Tyr Gly Ala Glu
305 310 315 320
Glu Cys Val Leu Gln Met Gly Gly Val Leu Cys Pro Arg Pro Gly Cys
325 330 335
Gly Ala Gly Leu Leu Pro Glu Gln Gly Gln Arg Lys Val Thr Cys Glu
340 345 350
Gly Gly Asn Gly Leu Gly Cys Gly Phe Val Phe Cys Arg Asp Cys Lys
355 360 365
Glu Ala Tyr His Glu Gly Asp Cys Asp Ser Leu Leu Glu Pro Ser Gly
370 375 380
Ala Thr Ser Gln Ala Tyr Arg Val Asp Lys Arg Ala Ala Glu Gln Ala
385 390 395 400
Arg Trp Glu Glu Ala Ser Lys Glu Thr Ile Lys Lys Thr Thr Lys Pro
405 410 415
Cys Pro Arg Cys Asn Val Pro Ile Glu Lys Asn Gly Gly Cys Met His
420 425 430
Met Lys Cys Pro Gln Pro Gln Cys Lys Leu Glu Trp Cys Trp Asn Cys

435

Gly Cys Glu

450

440

445